Tetralogy of Fallot with Unilateral Pulmonary Atresia
A Clinically Diagnosable and Surgically Significant Variant

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Four patients with tetralogy of Fallot syndrome, whose physical and x-ray findings were distinctly unusual, are presented in this report. All these patients were demonstrated to have functional or anatomic atresia of one of the main pulmonary arteries, in addition to the characteristic features of the tetralogy of Fallot. Diagnostic criteria, including angiograms, are presented, and the therapeutic implications are discussed.

Since the report by Blalock and Taussig in 1945 of the first successful "shunt operations" in three children with the tetralogy of Fallot syndrome, well over 1,000 such patients have been subjected to surgery throughout the United States.1,2

The successful accomplishment of an aortopulmonary shunt in a patient with tetralogy of Fallot will depend to a large extent on the availability of a pulmonary artery through which systemic blood can be directed to the lungs and on the presence of a second vessel to maintain the pulmonary circulation during the period of temporary occlusion of the first pulmonary vessel.

In the majority of patients with tetralogy of Fallot, the diagnosis is made on clinical grounds alone. The history, physical examination, roentgenologic findings, and the electrocardiogram in the typical case represent a relatively easily identifiable entity. As has been pointed out by Taussig and others, the presence and the size of the pulmonary arteries in the individual case may be quite difficult to assess clinically as well as by angiography.

The purpose of this paper is to report four patients with tetralogy of Fallot in whom the unusual distribution of the pulmonary arteries makes surgical correction a very hazardous undertaking. Physical and radiologic signs make this a well recognizable clinical entity.

Case Reports

The individual case reports are given below, with the salient points summarized in table 1.

Case 1. J. D. (no. 356134) was the product of an uneventful pregnancy and delivery. Cyanosis and labored respirations appeared six hours after birth and persisted despite administration of oxygen. Feedings were taken poorly, and frequent suctioning of the oropharynx was necessary because of excessive mucus.

Physical examination revealed a moderately cyanotic male infant in no acute distress. Temperature was 99.4 F., pulse rate 116, respirations 48 per minute, and blood pressure 84 (systolic). There was no clubbing of the fingers, deformity of the chest, distention of neck veins, or peripheral edema. Femoral and radial pulses were equal and strong. The liver extended 2 cm. below the right costal margin in the nipple line. No thrills were felt. A grade IV, rough systolic murmur was heard all over both sides of the chest. The murmur was loudest over the right upper chest anteriorly.

The hemoglobin was 17 Gm. per 100 ml., the erythrocyte count 5.6 million per cubic millimeter, and the hematocrit 52 per cent. The electrocardiogram showed right axis deviation and right ventricular hypertrophy.

Films and fluoroscopic examination of the chest revealed a considerable shift of the heart and mediastinum to the left, obscuring most of the left lung. The right upper and middle lobes appeared emphysematous, and the right lower lobe was collapsed. The right hilum contained a large conglomerate area of increased density, some portions of which were rounded and suggested a
vascular structure. The heart was not significantly enlarged, but the right ventricle was prominent.

A bronchogram, taken because of the evidence of obstruction of air exchange to the right lung, showed normal bronchi on the left, but flow of the opaque medium into the right lung was almost completely obstructed at the bifurcation of the right main bronchus by the rounded hilar densities previously described (fig. 1). These hilar masses appeared on angiography to be vascular (fig. 2), suggesting the possibility of an arteriovenous malformation in this area. No left pulmonary artery could be demonstrated on the films. There was overriding of the aorta with a right aortic arch, and the thoracic aorta descended on the right.

Table I.—Summary of Important Clinical and Laboratory Findings in Four Cases of Tetralogy of Fallot with Atrioventricular Canal

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age</th>
<th>Cyanosis</th>
<th>P2*</th>
<th>Maximal Murmur</th>
<th>X-Ray Findings</th>
<th>Confirmed by</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Yrs.</td>
<td></td>
<td></td>
<td></td>
<td>Pulmonary Artery</td>
<td>Pulmonary Vascularity</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Left</td>
<td>Right</td>
</tr>
<tr>
<td>1</td>
<td>77</td>
<td>Yes</td>
<td>?</td>
<td>Systolic</td>
<td>Enlarged</td>
<td>Absent</td>
</tr>
<tr>
<td>2</td>
<td>43</td>
<td>Yes</td>
<td></td>
<td>Systolic</td>
<td>Enlarged</td>
<td>Atretic</td>
</tr>
<tr>
<td>3</td>
<td>34</td>
<td>Yes</td>
<td></td>
<td>Systolic</td>
<td>Enlarged</td>
<td>Atretic</td>
</tr>
<tr>
<td>4</td>
<td>33</td>
<td>Yes</td>
<td></td>
<td>Systolic</td>
<td>Enlarged</td>
<td>Large</td>
</tr>
</tbody>
</table>

* Pulmonic second sound.
† Second right intercostal space.
‡ Right ventricular hypertrophy.
§ Right ventricular hypotrophy.

Although it was felt that the patient suffered from congenital heart disease, the possibility of an arteriovenous aneurysm, in addition, compressing the right main bronchus, seemed to warrant a right exploratory thoracotomy. At operation, an aneurysmal mass was present in the right hilum. A thrill was felt throughout the right lung and especially strongly over the hilar mass. This thrill disappeared on compression of the hilum. Such compression, however, caused a 10 to 20 per cent drop in blood oxygen saturation as indicated by an oximeter placed on the patient's ear. It was decided that the baby probably did not have sufficient oxygen uptake from his left lung to survive a right pneumonectomy. Consequently, the chest was closed.

Following surgery, respirations became increasingly rapid and labored, and cyanosis increased. Death occurred on the second postoperative day.

At autopsy, the heart was slightly enlarged, and the right ventricle was hypertrophied. The pulmonic ring was formed by a tough band of tissue only 1.6 cm. in circumference, and from this the orifice may be calculated to be 4 mm. or less in diameter. No pulmonic cusps were present. In their stead were several tiny greyish-white granular elevations. The pulmonary artery passed directly to the right hilum without there being any left pulmonary artery at all. In the right hilum the pulmonary artery rapidly dilated into a thin-walled sac which measured 3.2 cm. in circumference. Several branches passed from this aneurysmal right pulmonary artery into the right lung. The only blood supply to the left lung was a tiny branch of the innominate artery which passed into this lung just above the hilum. A high ventricular septal defect, measuring 0.7 cm. in diameter, was present. The aorta directly overrode this defect, and there was a right aortic arch and right descend-
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and blood pressure 92/52 (left arm). There was mild cyanosis of the lips, fingers, and toes, without clubbing. The neck veins were not distended, and there was no chest deformity or peripheral edema.

Femoral pulsations were strong. The liver extended 4 cm. below the right costal margin in the nipple line. The apex was in the fifth left intercostal space, 0.5 cm. beyond the midclavicular line. No thrills were felt. The second sound was diminished in the pulmonic area but was quite loud along the lower left sternal border. A grade III, rough systolic murmur was best heard in the third left intercostal space near the sternum. There was a second harsh systolic murmur, grade IV in intensity, loudest along the upper right sternal border. This murmur radiated well to all of the right chest as well as to the right side of the neck.

Laboratory studies showed a hemoglobin of 14 Gm., an erythrocyte count of 8.0 million, and a hematocrit of 47 per cent. Circulation time, arm to tongue (fluorescein), was 8 seconds. The electrocardiogram showed right axis deviation and marked right ventricular hypertrophy.

Films and fluoroscopic examination of the chest showed a slight increase in the transverse diameter of the heart with definite enlargement of the right ventricle. The normal pulsation of the main pulmonary artery was not seen at fluoroscopy, but there was no concavity in the left upper portion of the cardiac silhouette. The vascular markings in the right lung were considerably increased and showed an active expansile pulsation. In contrast, the vascular markings in the left lung were definitely diminished in caliber. There was a left aortic arch.

An angiocardiogram showed simultaneous filling of the pulmonary artery and the aorta from the right ventricle. There was stenosis of the left pulmonary artery in the region of its bifurcation with very little, if any, contrast medium being visualized in the peripheral vessels of the left lung. The right pulmonary artery was dilated, and its peripheral branches were unusually large (fig. 4a). The left atrium filled by pulmonary veins leading from the right lung, with no vessels from the left lung being seen. The aorta was refilled from the left ventricle, proving the presence of overriding (fig. 4b).

At cardiac catheterization, the pressure in the pulmonary artery and its right branch was 18/7,
of 83 per cent, a right-to-left shunt of 1.6 liters per square meter of body surface area per minute, and a left-to-right shunt of 0.7 liters per square meter of surface area per minute.

Case 3. R. S. (no. 341910). This 3½ year old white boy was the result of an uneventful pregnancy except that the mother had measles during the second month. The baby was cyanotic at birth, and a loud systolic murmur was heard at that time. Squatting, dyspnea on exertion, and increasing cyanosis appeared after the patient began to walk. At the time of admission, he could walk only two blocks and could climb one flight of stairs with great difficulty. There was no history of epistaxis, hemoptysis, spells, or congestive cardiac failure.

On physical examination, the patient was well developed and nourished and showed moderate cyanosis and clubbing. Temperature was 99.2°F., pulse rate 100, respirations 20 per minute, and blood pressure 110/80 (left arm). There was no peripheral edema, distention of neck veins, or chest deformity. Femoral and radial pulses were equal and strong. The liver extended 3 cm. below the right costal margin in the nipple line. The apex was in the fifth left intercostal space, 1.5 cm. beyond the midclavicular line. No thrills were present. The first sound was normal, but the second sound was very loud and single in the region of the third left intercostal space near the sternum. There was a rough, grade IV, systolic murmur which was loudest in the second and third right intercostal spaces near the sternum (fig. 5). This murmur was transmitted to the entire right chest and to the precordium.

The hemoglobin was 21.0 Gm., the erythrocyte count 8.0 million, and the hematocrit 69 per cent. Circulation time, arm to tongue (fluorescein), was 8

Fig. 3. Schematic presentation of autopsy findings in case 1. Note overriding aorta, ventricular septal defect, and pulmonic stenosis with large right main pulmonary artery. Left main pulmonary artery is absent.

Fig. 4a. Chest film of case 2. Note the engorged vascular pattern on the right.
seconds. The electrocardiogram showed right axis deviation, atrial hypertrophy, and marked right ventricular hypertrophy.

fig. 4b. Angiocardiogram of case 2. Note large right main and absent left main pulmonary artery.

fig. 5. Phonocardiogram of case 3. Note the diamond-shaped systolic murmur which is more intense on the right than on the left.

Films and fluoroscopic examination of the chest showed the heart to be slightly enlarged with definite prominence of the right ventricle. Normal pulmonary artery pulsations were absent along the upper left cardiac border; however, no concavity was seen in this area. The branches of the right pulmonary artery were markedly engorged, whereas the vascular markings in the left lung were strikingly diminished (fig. 6). The aortic arch was on the left.

An angiocardiogram showed simultaneous filling of a dilated right pulmonary artery and the aorta (fig. 7). The left pulmonary artery and its branches were not visualized, indicating absence, atresia, or marked stenosis of this vessel. Later films showed the aorta being refilled from the left ventricle.

A cardiac catheter was passed through the right atrium and right ventricle into the aorta. Despite
repeated efforts, it was impossible to pass the catheter into the pulmonary artery. Right ventricular pressure was 85/5. The oxygen saturation in the femoral artery was 70 per cent, and there was a right-to-left shunt of 6.4 liters per square meter of body surface area per minute.

Films and fluoroscopic examination of the chest confirmed the clinical and electrocardiographic impressions of dextrocardia and situs inversus. There was slight cardiac enlargement with prominence of the transposed right ventricle. The right pulmonary vascular markings were markedly decreased in number and caliber, while the vascular markings on the left were prominent and abundant (fig. 8). There was a right aortic arch with a right descending aorta.

An angiocardiogram showed simultaneous early filling of the aorta and the left pulmonary artery (fig. 9). No contrast medium was seen in the right lung until much later when a very small amount appeared, apparently reaching the right lung from a small branch of the aorta.

DISCUSSION

The tetralogy of Fallot syndrome consists of pulmonic stenosis, right ventricular hypertrophy, ventricular septal defect, and dextro-position of the aorta. The pulmonic stenosis encountered in this condition may be sub-valvular or valvular in location. The pulmonary artery and its left and right main branches show varying degrees of hypoplasia, which is classically symmetric and probably roughly proportionate to the stenosis of the outflow tract. The present group of patients can be set apart from the usual picture of the tetralogy of Fallot cases by the fact that, in addition to the stenosis of the valvular or sub-valvular area, they all showed atresia (functional or anatomic) of one main pulmonary artery with dilatation of the other.

The only detailed pathologic report in the literature of a tetralogy of Fallot patient with absence of one main pulmonary artery is that of Thomas in 1941. Blalock states that out of 610 patients with cyanotic congenital heart disease, he has encountered nine cases with absence of one pulmonary artery. He does not state the relative frequency with which each of the two sides was involved, but the two patients he has used as illustrations in this and another publication show the left main pulmonary artery to be absent.

It seems more than coincidental that all three of these documented cases and all three of our patients with the heart in normal position showed atresia of the left main pulmonary artery with dilatation of the right main vessel, especially since our one case with dextrocardia is the only one to our knowledge in which atresia of the right pulmonary artery is accompanied by dilatation of the left.

This correlation of atresia of the left main pulmonary artery with the tetralogy of Fallot syndrome becomes even more interesting when we consider the fact that all the reported instances of atresia of one pulmonary artery without intrinsic heart disease affected the right pulmonary artery. We have one such patient under observation at present at the Children's Medical Center in Boston.

To the best of our knowledge, no clinical criteria have so far been outlined to differentiate the present group of patients from the conventional type of tetralogy of Fallot cases. We believe, on the basis of the findings in our four patients, that such a differential diagnosis is possible on the following grounds:

(a) The systolic murmur of tetralogy of Fallot, if present, is always best heard at the left sternal border and transmits best to the left chest and the left clavicle. Our three patients with the heart in normal position demonstrated systolic murmurs with maximal intensity in the right chest and under the right clavicle.

Patients with tetralogy of Fallot and dex-
trocardia usually demonstrate systolic murmurs best audible over the right chest and under the right clavicle, whereas our one patient with dextrocardia had a murmur much louder under the left clavicle.

These auscultatory phenomena may well be due to the propagation of the murmur in the direction of the pulmonary blood flow

(b) The roentgenologic examination of patients with tetralogy of Fallot almost invariably shows a symmetric diminution of the pulmonary vascular pattern in the lung fields. In the cases with tetralogy of Fallot and unilateral pulmonary atresia, the affected side shows marked diminution of the pulmonary vasculature, whereas the side with the dilated main pulmonary artery demonstrated evidences of pulmonary vascular engorgement, occasionally even with a "hilar dance."

(c) Angiocardiographic studies confirm the presence of this entity by demonstrating an overriding aorta, atresia of one main pulmonary artery, and aneurysmal dilatation of the other.

(d) Cardiac catheterization studies demonstrate the presence of pulmonary stenosis. It is interesting to note that in two of our patients, catheterization strongly suggested the presence of a valvular stenosis—a finding also present in our one autopsied case.

Discussing the operative hazards in patients with a single pulmonary artery, Blalock\[6\] makes the statement that the risk is much higher in these cases than in patients with the conventional tetralogy of Fallot. The danger occurs at the time in the procedure when it is necessary to occlude completely the only pulmonary artery available, thereby stopping the entire pulmonary circulation, save for the blood reaching the lungs through collaterals. The danger inherent in this set of circumstances was well demonstrated during the operation on case 1 when momentary compression of the right pulmonary artery resulted in a rapid and significant drop in arterial oxygen saturation.

Although a shunt procedure may have to be attempted in a severe case of tetralogy of Fallot with unilateral pulmonary atresia, the indications for operation should clearly be much more stringent than under ordinary circumstances.

One may speculate on the feasibility of attempting a Brock procedure in these patients, but the autopsy findings of absent semilunar cusps in our patient and in Thomas' case make one hesitate to make such a recommendation, except as a last resort.

SUMMARY

1. Three patients with tetralogy of Fallot and atresia of the left main pulmonary artery are reported. All three have shown an associated dilatation of the right main pulmonary artery with pulmonary vascular engorgement on this side.

2. One patient with dextrocardia and tetralogy of Fallot showed atresia of the right main pulmonary artery with dilatation of the left.

3. Clinical and roentgenologic points in the differential diagnosis of these conditions are stressed.

4. The increased operative hazards represented by these patients are discussed.

SUMARIO Español

Se presentan en este informe cuatro pacientes con el síndrome de la tetralogía de Fallot, cuyos hallazgos físicos y radiológicos fueron distintamente poco usuales. En todos estos pacientes se demostró una atresia funcional o anatómica de una de las arterias pulmonares en adición a los hallazgos característicos de la tetralogía de Fallot. Criterio diagnóstico, incluyendo angiocardiogramas, se presentan; y las inferencias terapéuticas se discuten.

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